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# Untreated Early Onset Scoliosis - The Natural Progression of a Debilitating and Ultimately Deadly Disease

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## 1. Introduction

An early onset idiopathic scoliosis refers to idiopathic deformity that presents itself from birth to ten years of age and includes an infantile type (identified from birth to three years of age) and a juvenile type (identified from three to ten years of age). In contrast to adolescent idiopathic scoliosis (identified from ten years of age to the age of skeletal maturity), the juvenile idiopathic scoliosis patients, especially male, have a much higher risk of large curve development, since deformity occurs at a younger age and thus has a longer growth potential. Males are on average diagnosed earlier and become skeletally mature at a later age than females (Lenke & Dobbs, 2007). While some juvenile curves spontaneously regress, approximately 70% of them progress and ultimately require some form of treatment, either observation, orthotic management or surgical correction, with each treatment form being used at different Cobb angle intervals and progression landmarks. The absence of treatment for progressive early onset scoliosis usually results in severe adult idiopathic scoliosis, a condition that reduces quality of life, endangers general health and may ultimately lead to serious cardiopulmonary complications and premature death.

In the following text, describing and referencing a case of an adult patient with untreated early onset idiopathic scoliosis, I will try to summarise evidence-based facts and data regarding health related issues, natural progression and surgical management of untreated early onset and adolescent idiopathic scoliosis that results in severe adult deformity.

## 2. Living with spinal deformity - a lonely man's life story

A male was born in 1958 at a maternity ward with normal labour and delivery to a family of five, his father working as a farmer and mother as a housewife. At the age of eight his parents noticed his bad posture with a hump on his back and the referring paediatrician diagnosed him with idiopathic scoliosis. At the time of his first admission to an orthopaedic hospital's paediatric ward in 1966, the records show that his health status and chest radiograph of heart and lung were normal and that skeletal radiographs showed a right thoracic primary curve with a Cobb angle measuring 65° and severe vertebral torsion between the fourth and tenth thoracic vertebrae. The thoracic curve was flexible and corrected under Glisson traction harness to 36°. He was treated with scoliosis exercises and

hard bed tractions and extensions daily as was customary at the time. After six months of treatment his thoracic curve Cobb angle measured 60° with curve correction under Glisson traction harness measuring 40°. After eight months of treatment he was discharged from the hospital and further treatment was to be continued after three months, yet records show no acceptance at the planned date. The reason for his non-attendance at follow-up was given by him during a later interview in which he described his mother's decision to stop his treatment due to difficult family circumstances after his father's death during his hospitalisation.

There is little medical history for this patient until the age of 51. He worked full-time as a furniture technologist and lived with his brother in an apartment block. In 2009 he was admitted to a general hospital's gastroenterological department because of three day duration melaena, resulting in hypocalcaemia and haemodynamically relevant normocytic anaemia. During hospitalisation, signs of chronic alcohol abuse with elevated liver enzymes, liver steatosis and predelirant syndrome became apparent. Oesophagogastroduodenoscopy revealed signs of reflux oesophagitis grade B, erosive antral gastritis and erosive bulbar duodenitis with a Forrest III duodenal ulcer, and antibiotic treatment was administered due to signs of acute bronchitis. In 2010 he visited an orthopaedic surgeon for back pain, originating from the left side of the thoracic spine and radiating to the area of posterior left hemithorax and left lumbar. He also had left hip and neck pain, and he complained about a four year duration of shortness of breath when walking short distances. Upon examination we found a short stature with obesity and severe kyphoscoliotic right thoracic deformity with left lumbar compensatory curve and normal neurologic status. His right thoracic rigid primary curve Cobb angle between Th4 and Th11 was 125° with the apex on Th8. There was also a kyphotic angulation with a Cobb angle of 70° between Th2 and Th10 with the apex on Th7 and a left lumbar compensatory scoliotic, slightly flexible curve with a Cobb angle of 75° between Th12 and L4 with the apex on the L2/L3 intervertebral disc. The deformity was classified according to the Lenke Classification System as type 3C+. Physical and radiological examination by a referred internal medicine physician revealed a generalised cyanosis, peripheral oedema, left basal lung wheezes and crackles, diminished right hemithoracic movements and breath sounds over the right lung field. Chest radiograph revealed a left shifted cardiac dilatation with signs of increased pulmonary vascular resistance on bilateral basal areas and the left lung field. Spirometric pulmonary function tests revealed a severe chronic combined obstructive-restrictive ventilatory disturbance, his ECG showed signs of right ventricular hypertrophy and blood laboratory tests revealed secondary polycythaemia with hypoxaemia, hypercapnia and compensated respiratory acidosis. Severe kyphoscoliotic deformity was found to be the cause of chronic respiratory hypoventilatory insufficiency, aggravated by a long-term smoking habit that resulted in right-sided decompensated heart failure. He was prescribed with bronchodilators, diuretics and home oxygen therapy.

Shortly after he was admitted to our spinal surgery department for further preoperative imaging work-up and respiratory physical therapy. His pulmonary function tests were as follows: forced vital capacity (FVC) 0,71 litres (21%), forced expiratory volume in one second (FEV1) 1,039 litres (14%) and FEV1/FVC index 54% with arterial oxygen saturation of 89%. Echocardiographic examination revealed a right ventricle and atrium hypertrophy with tricuspid regurgitation (Fig. 1.).

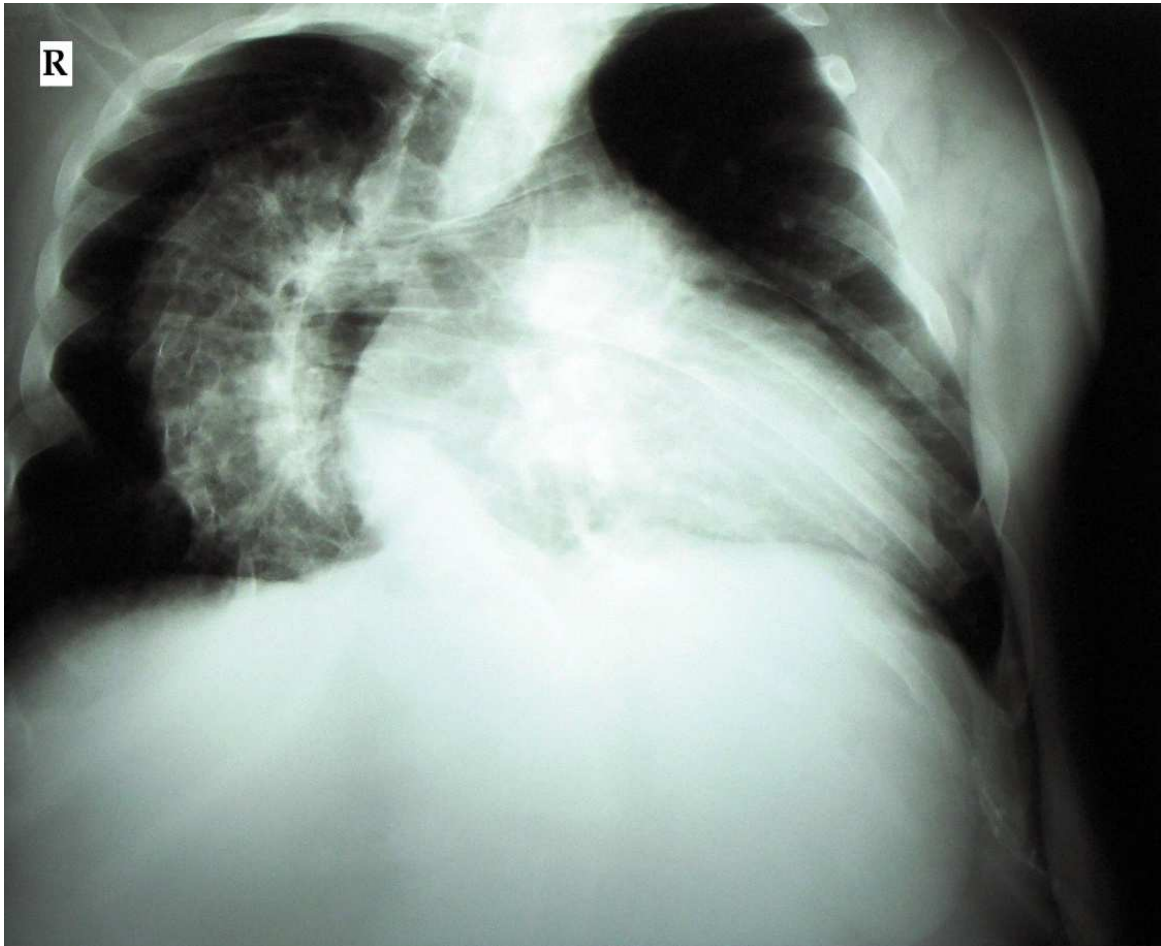


Fig. 1. Anteroposterior chest radiograph showing cardiac enlargement (cardiothoracic index measuring 60%) and severe right thoracic spinal column curvature.

The patient was scheduled for corrective deformity surgery but died at home in his sleep at the age of 52, shortly before his intended admittance to the hospital, due to a cardiorespiratory failure as a consequence of a severe combined obstructive-restrictive respiratory disease and pulmonary heart decompensation. No autopsy was performed.

### **3. Untreated idiopathic scoliosis - a spine surgeon's perspective**

An untreated idiopathic scoliosis after skeletal maturity is one of spine surgeon's greatest challenges, because its three dimensional rigid deformity and patient's often decreased physiological reserve related to cardiopulmonary insufficiency, chronic pain and age present a complex multidisciplinary approach to solving the problem, where doing as little as needed to achieve the best possible overall result often contradicts our way of surgical thinking, accustomed to successfully treating young and generally healthy adolescents (Fig. 2.).



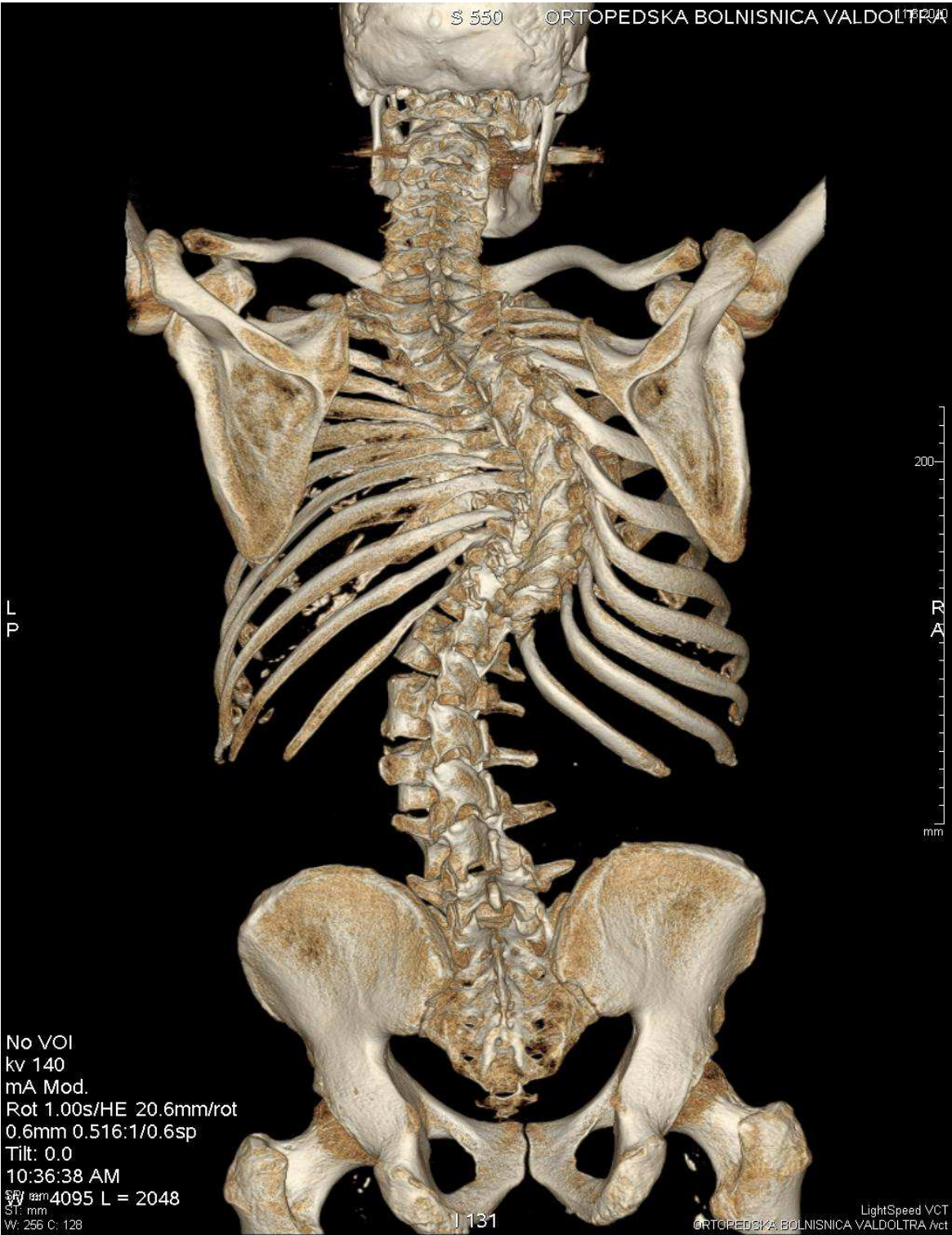


Fig. 2. 3D CT preoperative reconstruction of patient's deformity.

The decision on the proper method of surgical treatment is often based on personal experience with a demanding technique, awareness of conflicting evidence of complications and of short- to long-term results of these relatively rare spinal operations.

#### **4. Mortality**

An untreated idiopathic early onset scoliosis has a worse prognosis than late onset adolescent type as concluded in the long-term follow-up study of untreated idiopathic scoliosis patients (Nilsson & Lundgren, 1968; Pehrsson et al., 1992). Both studies report significantly increased mortality from respiratory failure or cardiovascular diseases compared to the general population with increased risk of death after 40 years of age. Increased mortality was found in patients with infantile and juvenile scoliosis, but not in patients with adolescent scoliosis, although it cannot be said that adolescent type never causes death from cardiopulmonary failure (Asher & Burton, 2006). A mortality of untreated scoliosis is related to the severity of the curve. In a study by Pehrsson et al. from 1991 it was found that respiratory failure occurred only in patients with a predicted vital capacity (VC) of less than 45% and a curve greater than 110°, the findings being consistent with the knowledge of the influence of deformity on immature lung development and function, that is before the termination of growth of the number of alveoli until the age of eight years (Pehrsson et al., 1991). In a study by Rom and Miller, the authors surprisingly encountered ten patients over a four year interval with kyphoscoliotic curves greater than 100° who had survived into the seventh decade without clinically significant cardiorespiratory embarrassment, although they concluded that severe deformity is not compatible with a long and active life (Rom & Miller, 1978).

##### **4.1 Health related issues**

There is a number of lifelong health impairment issues related to the untreated adult idiopathic scoliosis. In a long-term follow-up and prognosis study of untreated patients suffering from idiopathic scoliosis, the authors analysed 194 patients who were seen at the University of Iowa between 1932 and 1948. In 69 of those patients pulmonary function studies were done with findings published in 1981 (Weinstein et al., 1981). They reported a significant correlation between reduction in VC and FEV1, and increasing severity of the curve only in patients with thoracic curves, yet a marked reduction and significant limitation of pulmonary function in the non-smokers did not occur until thoracic curve approached a Cobb angle of 100°. In the later follow-up study of the same baseline group of patients the authors concluded that a Cobb angle greater than 50° at skeletal maturity is a significant predictor of decreased pulmonary function (Weinstein et al., 2003). Shortness of breath seems to be the main symptom of decreased pulmonary function in adult idiopathic scoliosis patients, associated with thoracic and large double curves with wheezing being the indicator of a poor prognosis. The new evidence shows that decreased chest wall motion, resulting in decreased pulmonary function, also affects adult patients with relatively small curves and that pulmonary stress testing should be the diagnostic method of choice. The mechanics of breathing in patients with severe thoracic scoliosis is similar to that seen in normal subjects with increased ventilatory drive and in patients with advanced chronic obstructive pulmonary disease with a pattern of respiratory muscle activation being essentially an automatic response of the central respiratory controller (Estenne et al., 1998).

A restrictive ventilatory impairment, a reduction in dynamic pulmonary compliance with breathing against an increased load and an impaired diaphragmatic function in patients with severe thoracic scoliosis, results in a rapid and shallow breathing pattern with increased activation of abdominal muscles during the expiration phase that results in a higher end expiratory gastric pressure, which is associated more frequently with symptoms of a gastroesophageal reflux disease (Fig. 3.).

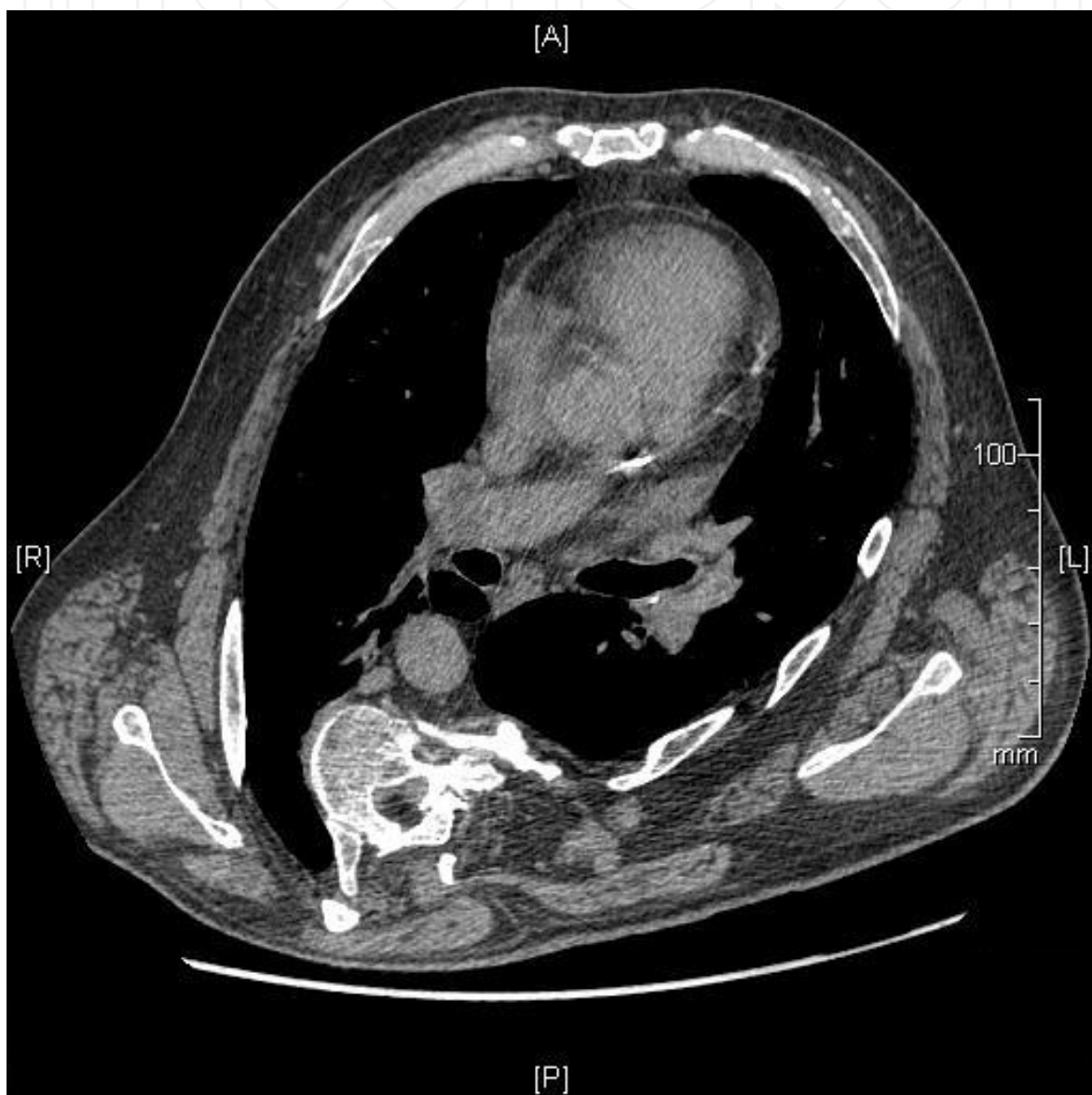


Fig. 3. Transthoracic axial CT image of a patient showing a marked rotation of a dysplastic thoracic vertebra, hypertrophic heart and decreased inflation of lung fields, occupying a reduced thoracic space.



The pathophysiological mechanism of respiratory impairment in spinal deformity follows a specific pattern; chest wall restriction results in a decreased VC and total lung capacity, tidal volume and flow rates are reduced in proportion to the reduction of VC. The ventilation-perfusion mismatch leads to hypoxaemia, resulting in active pulmonary vasoconstriction and hypercapnia secondary to alveolar hypoventilation with renal compensation of respiratory acidosis. Pulmonary vasoconstriction and pulmonary hypertension lead to right ventricular hypertrophy (cor pulmonale), ultimately resulting in right heart failure. An acute respiratory failure that usually follows an acute respiratory infection and right heart failure are thus the two most frequent causes of death in patients with untreated severe kyphoscoliotic deformity.

An increased risk of cardiovascular diseases has been reported among the population of untreated idiopathic scoliosis patients with systemic arterial hypertension being the most commonly overrepresented finding. There seems to be the same pathophysiological mechanism linkage of developing arterial hypertension among patients with obstructive sleep apnoea and night time hypoventilatory episodes with desaturations occurring in patients with severe scoliosis. There is also a higher incidence of congenital heart valve disease among the adult scoliotic population.

The prevalence of both acute and chronic back pain is more common among untreated scoliosis patients than among the general population, with symptoms rarely causing disability. The typical adult scoliotic patient's back pain is rarely severe, usually asymmetrical, often extravertebral and almost always disappears with rest. A long-term pain management programme is rarely needed. There seems to be no correlation between the type and the severity of the curve and back pain, except among patients with thoracolumbar curves, where a translatory shift of the vertebrae has been postulated as the cause, although Kostuik and Bentivoglio found an increasing prevalence and severity of pain with curves greater than 45° (Kostuik & Bentivoglio, 1981, as cited in Hu, 2006). It seems that double curves are least likely to be associated with increased pain. The more frequently reported site of back pain is on the convexity of the curve and in female patients the onset of back pain can be related to the first pregnancy. Lumbar radiculopathy can occur and appears to be confined to the concave side of the compensatory curves but other neurologic impairment is rare (Fig. 4.).

The knowledge of the impact of deformity on quality of life among adult patients with untreated idiopathic scoliosis is somewhat incomplete. Self-image is decreased, but less prevalent than among adolescent patients with idiopathic scoliosis. The frequency of mental health problems does not differ from the general population, although Ascani et al. in their uncontrolled analysis of a group of 187 patients with untreated idiopathic scoliosis and an average follow-up of 33.5 years found real psychological disturbances in 19% of cases, mostly women with thoracic curves greater than 40° (Ascani et al., 1986). Recent studies have showed that marriage and childbearing rates are not affected by adult idiopathic scoliosis to the threshold of relatively larger curves, although the Stockholm series in 1968 found 76% of female patients were unmarried. The authors of an Italian series study from 1986 concluded that the highest percentage of married patients was among those with thoracolumbar curves and the lowest percentage of unmarried patients was among women with thoracic curves. Adult idiopathic scoliosis does not affect employment rate or inability to work, although women with curves greater than 40° and patients with lumbar curves adapt less well to strenuous physical work.



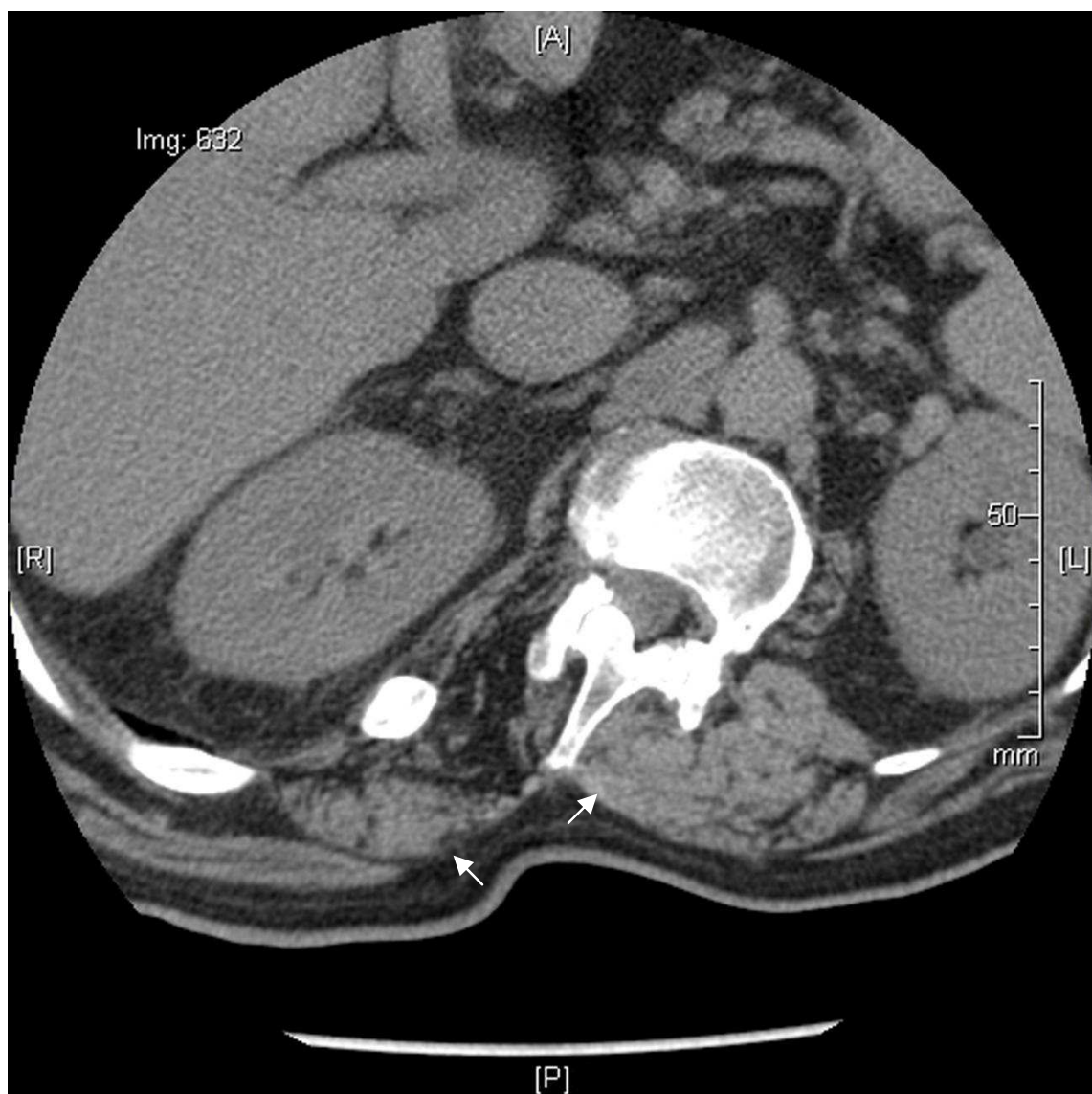


Fig. 4. Transabdominal axial CT image of a patient: notice a shift in abdominal organs due to vertebral rotation and asymmetry of paravertebral muscles (arrows).

#### 4.2 Natural progression of deformity in adult idiopathic scoliosis

The progression of idiopathic scoliotic deformity after skeletal maturity depends on the type and severity of the curve. The threshold for progression seems to be a Cobb angle of  $30^\circ$ . Single thoracic curves between  $50^\circ$  and  $80^\circ$  are most likely to progress at an average of  $0.73^\circ/\text{year}$  with an additional risk factor for progression being the apical vertebral rotation of more than 30%. Another risk factor for progression is a translatory shift between two vertebral segments that usually takes place at the lower end of the curve or at the transitional vertebrae, most frequently in the thoracolumbar curves. The lumbar component

of double major curves is more likely to progress than the thoracic component and right lumbar apex curves are twice as likely to progress than left apex lumbar curves. The lack of L5 deep seating and greater than 33% apical rotation in lumbar curves are also risk factors for progression. Table 1. shows a comparison between two groups of patients with untreated adult idiopathic scoliosis with regard to curve progression after skeletal maturity based on studies of by Weinstein et al. (2003) and Ascani et al. (1986).

			Iowa study group	Italian study group
No. of patients			117 (79 available for curve progression analysis)	187
Mean age			66 years	43 years, 7 months
Sex	Male		13 (11%)	36 (19.2%)
	Female		104 (89%)	151 (80.8%)
Average follow-up			51 years	33.5 years
Curve type	Thoracic		48 (41%)	74 (39.6%)
	Thoracolumbar		14 (12%)	29 (15.5%)
	Lumbar		32 (27%)	17 (9%)
	Double major		23 (20%)	67 (35.9%)
Average progression of curve by type	Thoracic		0.47° /year	0.47° /year
	Thoracolumbar		0.90° /year	0.40° /year
	Lumbar		0.28° /year	0.56° /year
	Double major	Thoracic component	0.26° /year	0.45° /year
		Lumbar component	0.31° /year	0.47° /year
Average progression of all types of curves combined			0.44° /year	0.47° /year
			0.46° /year	

Table 1. A comparison of demographic and curve progression data between two groups of patients with untreated adult idiopathic scoliosis.

The average progression of all types of curves combined is the same for both groups of patients, regardless of an age difference of approximately 20 years between groups, the conclusion being that the progression of the curve after skeletal maturity shows the same steady increase. Based on relevant literature findings, it is safe to conclude that the progression of any type of curve in idiopathic scoliosis after skeletal maturity rarely exceeds 1°/year, although there may be rare exceptions to this rule. In contrast with the findings shown in Table 1, other relevant literature suggests that thoracic curves tend to increase more than lumbar curves, lumbar curves more than thoracolumbar curves and thoracolumbar curves more than double major curves. Curves in untreated scoliotic women who have had one or more pregnancies progress somewhat more than curves in women with no pregnancies. Patients with discernible body alignment decompensation at the end of growth seem to improve with time.

During the 43 years since his last measurement, the Cobb angle of thoracic curve of the reported patient progressed from 60° to 125°, the average progression being therefore 1.5°/year, although this rate is exaggerated since most of the progression of the curve must have happened whilst growing up. Since we do not have radiographs from 1967, the measurement was taken from the patient's written records - we cannot asses the time of his skeletal maturity and progression of the curve until then.

### **4.3 The development of early onset idiopathic scoliosis surgery - from Harrington hook and rod instrumentation to modern-day operative techniques**

In 1962 Dr. Paul R. Harrington published his results in treating idiopathic scoliosis by introducing a metal hook and rod system, and a major advance in the field of surgical management of spinal deformities was made. The original Harrington rod operated on a ratchet system, attached by hooks to the spine at the top and bottom of the curvature that, when cranked, would distract or straighten the curve. At the time of the reported patient's hospitalisation for his early onset idiopathic scoliosis, the corrective operations of spinal deformities with Harrington instrumentation were common in Slovenia, yet his termination of treatment and the lack of follow-up ruined his chances of leading a relatively healthy life. We can only speculate that the attending physician planned to continue conservative treatment with tractions and bracing until his adolescence, since the knowledge of the impact of spondylodesis on the growing spine (i.e. danger of developing a crankshaft phenomenon and loss of truncal height) was available at the time or that he would have been scheduled for repeated instrumented corrections until the right age for definitive fusion.

Long-term follow-up studies of patients, operated upon with Harrington instrumentation for juvenile and adolescent idiopathic scoliosis, report good general functioning compared with sex and age matched control groups. No significant differences in the single Scoliosis Research Society (SRS) Instrument domain scores between operated patients and control groups, no important impairment of health-related quality of life, no significant correlation between patient's self-reported outcome and preoperative curve magnitude or postoperative rate of correction and an overall high degree of long-term postoperative satisfaction were found (Dickson et al., 1990; Padua et al., 2001; Mariconda et al., 2005). Although Harrington instrumentation was historically a very successful system for treating juvenile and adolescent idiopathic scoliosis, some drawbacks and complications have been reported, namely some loss of correction after time, pseudoarthrosis and broken rods, no improvement of the rib hump with correction being achieved only in coronal plane, flat back syndrome and associated adjacent segment disease at the most caudad levels of fusion that may result in higher incidence of back pain and back fatigue. Despite these complications and the fact that differences in study designs do not allow for a direct comparison of long-term follow-up studies of untreated adult idiopathic scoliosis and studies of patients operated upon with Harrington instrumentation for idiopathic scoliosis, the reported patient would have undoubtedly led a very different life with regards to quality, health issues and life expectancy if he had been operated upon when young.

A preoperative plan for early onset idiopathic scoliosis has to include issues of expected postoperative loss of spinal height and limited growth of the thoracic cage, affecting lung development. The risk of developing a crankshaft phenomenon postoperatively can be prevented by the use of standard anterior and posterior fusion procedures, indicated for older juveniles or growing-rod posterior instrumentation systems, indicated for younger children in whom a diminished chest-wall growth and volume is a concern. The growing-rod posterior instrumentation systems include posterior growing single-rod and Luque trolley systems, dual growing-rod and hook/screw implant systems, McCarthy's Shilla technique of posterior apical growth arrest and fusion construct with proximal and distal non-locking screw to rod fixation and rib-to-rib or ribs-to-spine hybrid constructs, widely known for its use in surgical management of thoracic insufficiency syndromes.

For patients with a scoliotic Cobb angle of more than 80°, with or without thoracic or thoracolumbar kyphosis of more than 70°, a perioperative halo-gravity traction protocol is recommended: a six up to eight-pin halo frame is placed slightly below the equator of the skull just above the eyebrows and the cephalad portion of the earlobes under general anaesthesia with pins tightened to six to eight inch pounds of torque, the axial traction weight is applied in the upright position of the spine through the halo and gradually increased daily for two to twelve weeks with cranial nerve and extremity neurologic examinations performed every eight hours and radiographic improvement of deformity monitored weekly until 32% to 50% of body weight loading is reached (Rinella et al., 2005). The purpose of this protocol, which is particularly useful in patients with previous spine surgeries or intraspinal pathology, is a slow perioperative correction of severe and rigid deformities and improvement of sagittal and coronal alignment while at the same time maximization of cardiopulmonary function and nutritional status before a definitive anterior and/or posterior fusion is performed.

Growth modulation techniques are new procedures utilised for smaller deformities to avoid secondary manifestations of scoliosis. They include a convex intervertebral disc stapling technique, in which continued concave growth maintains or even corrects a deformity over time, and anterior tethering technique, in which a polypropylene tether, attached to the heads of the screws, placed in the vertebral bodies anteriorly, prevents motion of the spine only when bending away from the tether.

#### **4.4 Adult idiopathic scoliosis - rethinking good luck in spinal deformity surgery**

Surgical treatment of severe adult idiopathic scoliosis deformity always presents a challenge for spine surgeons, not just because of demands of technically difficult operations, but also because of a high-level mental processing during diagnostic and patient selection stages of treatment, postoperative addressing of frequent complications and a long rehabilitation process. As a rule, a multidisciplinary approach is used and the key to successful treatment outcome is proper patient selection, where the benefits of surgery outweigh the risks. The indications for surgery include: back pain failing conservative care, progressive leg pain or neurologic deficit, muscle fatigue secondary to spinal imbalance, curve progression, progressive pulmonary compromise secondary to deformity and severe deformity resulting in poor body image, with goals of surgery being: a decrease in pain, stability of the curve, cessation of progression of the curve and pulmonary compromise, and improvement of neurologic symptoms (Hu, 2006).

Adult idiopathic scoliosis surgery differs from adolescent idiopathic scoliosis in terms of higher rates of reported mortality and morbidity associated with complications. In its 2011 degenerative lumbar and major adult deformity analysis the SRS queried its morbidity and mortality multicentre, multisurgeon database for the year 2007, associated with 22,957 reported cases, 6,782 of them being adult scoliosis, and stratified it using the American Society of Anesthesiologists' (ASA) physical status classification (Fu et al., 2011). They found a 8.4% overall complication rate and complication rates for ASA Grades 1 through 5 being 5.4%, 9.0%, 14.4%, 20.3% and 50.0%, respectively with mortality rates being 0.03%, 0.1%, 0.3%, 2.2% and 33.3%, respectively. In the subgroup of adult patients with spinal deformity, morbidity and mortality rates for ASA Grades 1 through 4 were 10% and 0%, 16% and 0.2%, 22% and 0.7% and 31% and 4%, respectively. In its 2011 SRS Morbidity and Mortality of



Adult Scoliosis Surgery report, the same database was queried to identify 4,980 submitted cases of adult scoliosis, 2,425 of them being adult idiopathic scoliosis, from 2004 to 2007 in order to assess complication rates on the basis of type of scoliosis, age, use of osteotomy, revision surgery status and surgical approach (Sansur et al., 2011). The overall complication rate was 13.4% with the previously reported adolescent idiopathic scoliosis surgery complication rate being 5.7%, therefore supporting the notion that advanced age leads to increased risk for the development of complications. The most common complications were dural tears (2.9%), implant complications (1.6%), deep wound infections (1.5%), acute neurologic deficits (1.0%), superficial wound infections (0.9%), delayed neurologic deficits (0.5%), wound haematoma (0.4%), epidural haematoma (0.2%), pulmonary embolus (0.2%), deep venous thrombosis (0.2%) and sepsis (0.1%). Significantly higher complication rates were identified in osteotomies (particularly infections), revision surgery and combined anterior-posterior surgery with complication rates not being influenced by scoliosis type (degenerative vs. adult idiopathic scoliosis) or age (less than 60 years vs. greater than 60 years). The mortality rate was 0.3% associated with cardiopulmonary causes, sepsis and excessive blood loss with intraoperative death. The complication rates were greater in patients with higher ASA grades with infections, respiratory complications and haematomas demonstrating a 7-fold, an 8-fold and a 9-fold increase in rate, respectively when comparing ASA Grade 1 patients to ASA Grade 4 patients.

Surgical site infections seem to be one of the most frequent complications in adult spinal deformity surgery associated with long operation times that involve exposure at several levels and increased blood loss, resulting in higher morbidity, mortality and healthcare costs. A retrospective analysis of 830 adult patients with a mean age of 55.4 years who underwent surgery for kyphosis or scoliosis between 1996 and 2005 demonstrated a 5.5% overall surgical site infection rate (2% superficial and 3.5% deep to fascia) with risk factors being obesity and a history of prior surgical site infections warranting a separate drain in a subcutaneous layer and a modification in perioperative antibiotic prophylactic regimens (Pull ter Gunne et al., 2010). The increased subcutaneous fat layer in obesity increases retraction forces to provide exposure, leading to increased tissue necrosis while at the same time increased tissue thickness creates dead space between fascia and skin stitches. For combined anterior-posterior surgeries, staged procedures with exposure to two anaesthetics and a recovery period between two surgeries seem to have a higher infection rate than same-day procedures. Malnourishment in the recovery period is a particular risk factor for developing infections and other complications, therefore a total parenteral hyperalimentation and/or staging procedures over a longer period is warranted, allowing nutritional depletion recovery to baseline.

Other most frequently reported adult scoliosis surgery complications are residual pain (reported rates ranging from 5% to 15%), pseudoarthrosis with or without implant failure (reported rates ranging from 5% to 27%), progressive spinal decompensation, postoperative non-resolving visual loss, delayed postoperative paraplegia attributed to ischaemia of the spinal cord from postoperative hypovolaemia and mechanical tension of spinal blood vessels on the concavity of the curve, syndrome of inappropriate antidiuretic hormone hypersecretion, pancreatitis, superior mesenteric artery syndrome, ileus, pneumothorax, haemothorax, chylothorax, lung atelectasis, pneumonia, fat embolism and urinary tract infections.

With reported increasing rates of complications with age for adults undergoing scoliosis surgery, the surgeon's and patient's decision for operation must be based on evidence-based reported outcomes. In its 2011 risk-benefit assessment of surgery for adult scoliosis, the SRS retrospectively reviewed its prospective, multicentre spinal deformity database and queried 206 patients, 86% of them having a history of untreated adolescent idiopathic scoliosis, who completed the Oswestry Disability Index (ODI), Short Form-12 (SF-12), Scoliosis Research Society (SRS-22) and Numerical Rating Scale (NRS) for back and leg pain questionnaires at two year follow-up (Smith et al., 2011). Adult scoliotic patients were distributed among age groups of 22 to 44 years, 45 to 64 years and 65 to 85 years, having perioperative complication rates of 17%, 42% and 71%, respectively, with elderly patients having greater disability, worse health status and more severe back and leg pain at baseline than younger patients. Decompression, osteotomy and pelvic fixation were more commonly performed in the surgical treatment of older patients with operating room time, blood loss and length of hospital stay being all significantly greater in the same group. At two year follow-up there were significant improvements in ODI, SRS-22 and NRS for back and leg pain within each age group and significant improvements in SF-12 among those aged 45 to 64 and 65 to 85. The authors concluded that these data demonstrate the potential benefits of surgical treatment for adult scoliosis and suggest that the elderly, despite facing the greatest risk of complications, may stand to gain a disproportionately greater improvement in disability and pain with surgery. They also stressed that occurrence of major complications with adult deformity surgery does result in a modest impact on general health status and that within the confines of appropriate practice and in the absence of significant progressive neurologic deficits, the ultimate management decision should typically be guided by patient choice. The question remains whether this also holds true for patients with progressive cardiopulmonary compromise facing surgery.

With these data in mind a rough preoperative assessment of mortality, complication rate and surgical benefits for the reported patient can be made. The patient was ASA Grade 4, therefore he would have a 4% mortality and a 31% complication probability with a probability for infections, respiratory complications, haematoma and thromboembolic events being 8.2%, 3.2%, 0.9% and 1.1%, respectively. When stratified by patient age ( $\leq 60$  years), type of scoliosis (idiopathic), use of osteotomy (yes), revision status (no revision) and surgical approach (posterior only) the patient would have a complication probability of 9.9%, 13.3%, 9.9% and 10.3%, respectively. Being obese the patient would have a significant probability for postoperative wound infection and by quoting Smith et al., his age group would represent a 42% overall probability for operative complications with probability for minor and major complications being 27% and 15%, respectively, although two years after surgery the expected improvement of disability, back pain and health status would be statistically significant. Based on presented statistical probabilities, an accurate overall probability for mortality, morbidity and improvement cannot be drawn because of the use of different statistical methods in the studies and therefore these values should serve simply as rough guidelines for surgeons, patients and health insurance companies. Major reconstructive surgery of deformities cannot be seen and understood from a strictly mathematical and technical point of view, for the nature of our trade deals with so many dependent and independent factors leading to different circumstances and finally outcomes that "artistic form" in its basic meaning may be a better term when trying to comprehend the unpredictable aspects of complex surgery.

Obtaining a solid fusion by providing rigid internal fixation while at the same time correcting curves and achieving spinal balance in the coronal and sagittal plane are the main goals of adult deformity surgery. Surgical planning in adult idiopathic scoliosis must address technical problems of adjacent segment degeneration and junctional kyphosis at the levels proximal to instrumented fusion, osteopenic bone requiring multiple fixation levels with hybrid systems of transpedicular screws, segmental hooks and/or sublaminar wires, rigidity of the curve requiring release procedures and/or osteotomies with emphasis on priority of achieving and maintaining coronal and sagittal balance to curve correction.

The technical possibilities of modern day spinal surgery allow a surgeon to correct and maintain correction and spinal balance in the most difficult triplanar deformities. It is usually the impact of corrective surgery on possible neurologic deficits and direct or indirect iatrogenous visceral damage that limits the degree of correction, warranting complex anaesthesia with perioperative neurovascular monitoring during the operation and extensive postoperative intensive care management during the recovery period. The danger of poor general health deterioration during and after the extreme physical stresses of prolonged and very invasive surgery is also an issue that demands modification of an otherwise technically feasible surgical plan. The surgical plan must address the issues of approach, levels of fusion, fixation techniques with the use of proper implant and instrumentation selection, spinal column and rib cage release and osteotomy techniques, proper deformity manipulation techniques together with construct manoeuvring and the use of optimal bone grafting methods with efficient bailout and conversion procedures available during every stage of operation in case of complications.

Controversy exists around using combined anterior and posterior approaches or just posterior approaches in correcting rigid spinal deformities, but the current trend is the use of safer posterior approaches that still allow the option of using several different osteotomy techniques. Combined approaches are used when addressing a multiple level rigid deformity requiring anterior release, anterior corpectomy and structural grafting, especially in thoracolumbar kyphoscoliosis for re-establishment of lumbar lordosis. The use of an open anterior approach in a high-risk patient with pulmonary compromise has its drawback in decreasing pulmonary function. In a prospective study on pulmonary function before and after anterior spinal surgery in adult idiopathic scoliosis, the authors found a long-term decrease in mean FVC of 0.21 litres in all patients and a decrease in mean FVC of 0.31 litres in patients who underwent anterior surgery without posterior surgery in spite of mean Cobb angle correction of 31% (Wong et al., 1996). They concluded that a deleterious effect on lung function in a combined approach appears to be more important than a mechanical benefit of correcting a curve, that posterior spinal surgery alone tends to have a beneficial effect on pulmonary function and that an anterior alone or a combined approach should not be undertaken to improve pulmonary function.

When addressing an adult deformity through posterior approach, three different osteotomy techniques; Smith-Petersen osteotomy (SPO), Pedicle Subtraction Osteotomy (PSO) and posterior Vertebral Column Resection (pVCR) are commonly used (Doward & Lenke, 2010). SPO is a posterior column osteotomy that includes a posterior release through facetectomy and re-alignment of the sagittal and/or coronal plane deformity through posterior instrumentation compression with a mobile disc serving as a fulcrum. With SPO, the degree of kyphotic correction is in the range of 9.3° to 10.7° per level or 1°/mm of bone resected and

the technique, although providing the least correction of the three, can be used on multiple levels with reduction in operative time, blood loss, and risk of neurologic complications. A PSO or eggshell osteotomy involves a resection of the pedicles, decancellation of a wedge of the vertebral body via a transpedicular corridor and a posterior closing of the wedge through instrumented compression. The corrective power of a single PSO is roughly the amount of three SPOs, yet the average blood loss is nearly twice as much as three SPOs. Indications for a PSO include angular kyphosis, or if performed asymmetrically, angular scoliosis, significant sagittal imbalance and corrections at levels lacking anterior flexibility. The degree of kyphotic correction is on average of 32° at the operated level, making it a powerful single osteotomy technique, but with expected higher technical demands, operative times, blood loss and complication rates, especially neurologic deficits, than SPO. The most powerful and demanding of the three osteotomy techniques is pVCR, where after a posterior resection of the entire spinal level, the proximal and distal limbs are brought together and the curvature of the spine is corrected with the use of an interbody cage, acting as a fulcrum. The corrective power of a single pVCR can be as much as 100° in kyphoscoliotic cases. Indications for pVCR include sharp, angular kyphotic and/or scoliotic deformity in the thoracic and thoracolumbar spine, resection of hemivertebrae and intravertebral spinal tumours and shortening of the spine proximal to a tethered region. The prevalence of complications when using pVCR is significant with longer operative times and increased blood loss.

The aim of treatment in the reported high-risk patient was the retainment or at best improvement of pulmonary function with correction of kyphotic and, to a moderate degree, thoracic scoliotic curve while at the same time enabling a good body balance by correcting the somewhat flexible lumbar scoliotic curve. The preoperative plan consisted of a three week halo traction, respiratory physical therapy and hyperalimentation after which a neurovascular monitored, all posterior approach would be performed with laminar hook placement from Th1 to Th4 and pedicle screw insertion from Th9 to L4 vertebrae. The rib release on the apical concave side of the curve would be performed next, followed by an asymmetrical triple SPO at Th6/7, Th7/8 and Th8/9 motion segment. A concave side distraction, a convex side compression and a derotation manoeuvre over a contoured rod would follow after which a pelvic cancellous and posterior element cancellous and cortical autograft would be placed on decorticated laminae and a standard wound closure would complete the operation. The patient would be mobilised within 24 to 48 hours in an intensive care unit and a custom thoracolumbosacral orthosis would be fitted in the following days. He would receive respiratory and orthopaedic physical therapy once moved to the ward and would be discharged from the hospital when physically independent, with follow-up visits scheduled one, three and six months after discharge, yet the potential success of this kind of treatment plan would sadly never be known.

## 5. Conclusion

Early and late onset idiopathic scoliosis, although treatable either by observation and exercises, bracing or surgery may become a serious health concern if left untreated. In modern health care systems with proper screening programmes of school age children and adolescents, the probability for missed cases and development of adult idiopathic scoliosis is low. School screening is not only an instrument for early detection and decrease in the



number of patients who will eventually experience operative treatment, but it is also an indispensable tool for research on scoliosis aetiology (Grivas et al., 2007). There is great diversity in the policies for scoliosis screening worldwide and between different professional associations, e.g., The American Academy of Orthopedic Surgeons recommends screening girls at ages 11 and 13, and screening boys at 13 or 14 years of age and The American Academy of Pediatrics recommends scoliosis screening with the Adam's forward bending test at routine health visits at 10, 12, 14 and 16 years of age. Still, with rare patients not responding to follow-up visits, as was the case with the reported patient, migration of people from rural to urban areas and immigration of people from developing and underdeveloped countries as a result of globalisation, a spine surgeon must expect to see and treat difficult cases of adult idiopathic deformity.

Early onset idiopathic, compared to late onset scoliosis has a worse prognosis if left untreated because of larger curve development potential and increased mortality risk from cardiopulmonary failure, related to severity of the curve. Patients with untreated adult idiopathic scoliosis have a decreased pulmonary function, especially those with a Cobb angle greater than 50° at skeletal maturity. They have an increased risk of cardiovascular diseases and they have a higher frequency of back pain compared to the general population. Adult idiopathic scoliosis, with the exception of severe curves, does not affect marriage and childbearing, mental health and employment rate or inability to work. The progression of idiopathic scoliotic deformity after skeletal maturity shows a steady increase of less than 1°/year in the majority of cases and single thoracic curves between 50° and 80° are most likely to progress with body alignment decompensation at the end of growth improving with time.

Surgical treatment of severe adult idiopathic scoliosis deformity is technically demanding, has high rates of reported mortality and morbidity and should therefore be reserved only for experienced spine surgeons. The complication rates are strongly correlated to ASA grades, advancing age, osteotomies, revision surgery and combined anterior-posterior approaches, but possible benefits for surgical treatment are great, especially among the elderly. In a modern tertiary health care service setting with the use of multidisciplinary diagnostic and supportive treatment options, intraoperative neurovascular monitoring and safer posterior osteotomy and instrumentation techniques, the possibilities of reducing risk and increasing the success rates of these challenging procedures are improving.

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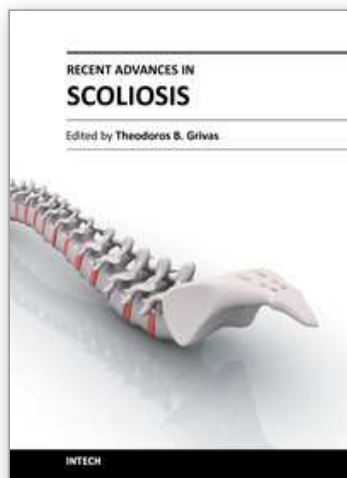
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## **Recent Advances in Scoliosis**

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This book contains information on recent advances in aetiology and pathogenesis of idiopathic scoliosis, for the assessment of this condition before treatment and during the follow-up, making a note of emerging technology and analytical techniques like virtual anatomy by 3-D MRI/CT, quantitative MRI and Moire Topography. Some new trends in conservative treatment and the long term outcome and complications of surgical treatment are described. Issues like health related quality of life, psychological aspects of scoliosis treatment and the very important "patient's perspective" are also discussed. Finally two chapters tapping the untreated early onset scoliosis and the congenital kyphoscoliosis due to hemivertebra are included. It must be emphasized that knowledgeable authors with their contributions share their experience and enthusiasm with peers interested in scoliosis.

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